ON HEREDITARY CHOREA

WITH A REPORT OF THREE ADDITIONAL CASES AND DETAILS OF AN AUTOPSY

BY

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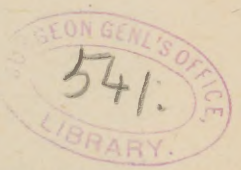

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ON HEREDITARY CHOREA, WITH A REPORT OF THREE ADDITIONAL CASES AND DETAILS OF AN AUTOPSY.[1]

THIS affection is one of considerable interest, partly on account of its rarity, but more especially from the peculiar and unvarying symptoms in every case which is met with.

The disease was first described by Dr. C. O. Waters, in a letter to Dr. Dunglison, which was published in his "Practice of Medicine," in 1841. It was also detailed by Dr. Charles B. Gorman, in a thesis for the degree of Doctor of Medicine, at the Jefferson Medical College, and referred to by Dr. Dunglison in the same volume of his "Practice." Dr. Irving W. Lyon reported some cases in the *American Medical Times*, in 1863, but it was not until Dr. Huntington, in 1872, read a paper on "Chorea," in which he mentioned the hereditary type, that the disease attracted any attention. From that time numerous cases have been reported, and the name of Huntington's chorea is frequently applied to the affection.

The choreic movements in this disease are much like those of St. Vitus's dance commonly met with in children, but they vary in certain peculiarities. They are more constant, more excessive, more rhythmical, and less under the influence of the will than in Sydenham's chorea. The patient is continually repeating the same irregular, jerky movements while sitting or standing, but when he attempts to walk the normal gait is interfered with by

[1] Read before the Medical Society of Virginia, October 8, 1891.

the grotesque and involuntary movements which are interposed. For example, as in the case of two of the patients, whom I shall describe later, when told to walk they would take a few steps quite naturally, then suddenly one leg would be violently thrust forward and the other brought up to it quickly with a little hop like a dancing step, then a few more sedate steps would be taken, to be again interrupted by the dancing step. Several writers have referred to this peculiarity in the gait.

In hereditary chorea, as the name implies, the disease is handed down from one generation to another; but we must bear in mind the distinction between hereditary chorea and the hereditary tendency to chorea. In the one the disease occurs in one generation after another, from father to son, or daughter, and following the same type; in the other, the disease presents itself in several children in the same family, differing perhaps in degree, and very frequently with no history of chorea in preceding generations, but instead, a history of some other neurosis. It is said that if the disease fails to appear in one generation all succeeding generations of this branch of the family are exempt from it. This, although generally the case, is not an unvarying rule.

The disease does not occur until middle life, beginning at about thirty-five years of age, and is essentially chronic. It never gets well, although there may be periods of relief, but, as a rule, it steadily goes on, getting worse until the death of the patient. This may not occur for many years, the disease lasting ten, fifteen, or even twenty-five years. Mental disturbances are common. Sometimes there are delusions of grandeur, and an elevated opinion of personal attraction, etc., and sometimes there is melancholia or dementia.

In September, 1888, I read before the American Neurological Association a paper [1] on "Hereditary Chorea," and recorded the histories of two new cases. Before that time only twelve different authors had reported cases,

[1] Journal of Nervous and Mental Diseases, February, 1889.

occurring in fifteen different families; but since then there have been extensive additions to the literature of the subject, and many cases have been reported in all parts of the world. A number of these cases have been found in insane asylums.

Klippel[1] relates a case of this affection in a man in whom there were irregular muscular movements beginning at the age of fifteen years. These increased in severity, but he remained in the army until the age of twenty-four years, when his condition was such that he was unfit for the service. The patient had always been irritable, and when he came under observation at the age of twenty-nine, he was morose, suicidal, and subject to attacks of violent rage. He was suspicious, and constantly thinking that someone was going to injure him. There was general chorea of all the muscles of the trunk, extremities, and face, and well-marked facial asymmetry.

Suckling[2] adds a case of hereditary chorea in a man of thirty-nine, who had been affected since about thirty-five years of age. The patient's mother died at fifty-six, having been choreic for sixteen years; and one of his sisters, aged thirty-eight, had had the disease for five years. Suckling states that a daughter of the patient, aged twelve years, was also choreic.

Theodore Diller, M.D., in a paper on this subject,[3] gives the histories of five cases; three of these were patients in the Iowa Hospital for the Insane, two of them being brothers. Another case was communicated to the author by Dr. G. Alder Blumer, under whose care he was in the State Lunatic Asylum, Utica, N. Y. The fifth case was under Dr. Diller, in the State Asylum for the Insane, Danville, Pa. This patient was one of a large family, ten in all; five of them were choreic.[4] Of the relatives of this patient, the father, two paternal aunts, and two

[1] Encéphale, vol. viii., p. 716. Paris, 1888.
[2] Birmingham Medical Review, September, 1889.
[3] American Journal of the Medical Sciences, December, 1889.
[4] I called attention, in the paper above referred to, to the fact that frequently these cases belong to large families.

paternal uncles had the disease. The grandfather was also choreic. The affection had begun at about the age of forty, and she had been affected mentally since the beginning of the chorea; she was melancholic and subject to fits of anger, in which she had threatened to poison her neighbors, and did attempt suicide. Diller, in this paper, expresses his doubts as to there being any difference in the essential characters of hereditary chorea and the ordinary chorea of children. He prefers to call them varieties of the same affection. It is true that the multiplication of the nomenclature of disease is undesirable; but the differences in all of the features of the two forms of chorea are so great that they are practically different affections. When the morbid anatomy of the disease has been studied sufficiently, no doubt there will be marked pathological differences found. W. P. Herringham, in an admirable paper on this subject,[1] expresses himself as follows: "The conclusion which comparison suggests is, that the two, that is, hereditary and Sydenham's chorea, are produced by affections of the same parts, but of a different nature."

In an interesting paper by Diller,[2] on "Chorea among the Insane," the writer records thirty-nine cases; of these thirty-three were furnished by the medical officers of twenty-three hospitals to which Dr. Diller had sent a circular asking for information on the subject. Of these there were but three cases of the hereditary form. These were reported by Dr. Diller in his paper already referred to. Dr. Diller's conclusions are that chorea is more common among the insane than is usually supposed. In the twenty-three asylums from which he got replies to his letters of inquiry, there are sixteen thousand four hundred and ninety-nine inmates; among these there were thirty-nine choreic, or a proportion of 1 to $423\frac{2}{3}$. I cannot agree with Diller that in all long-standing cases of chorea there is more or less marked tendency to mental deterio-

[1] Brain, October, 1888.
[2] American Journal of the Medical Sciences, April, 1890.

ration. There are many cases of chronic chorea in which the mental condition is normal, and I recall the case of a man of thirty-four, who had been choreic from birth, who was, as far as could be observed, normal mentally, and was able to support himself by the business which he carried on.

Bower[1] records three cases of this form of chorea. The first was in a negro, not a full-blooded African, however. The patient has almost straight hair and a copper-colored skin. His mother, her two sisters, and one brother had the same affection. The patient was thirty-six years of age, and the disease had begun at about thirty. He had been a hard drinker, and had a history of paraplegia, coming on when he was twenty eight and lasting about three months. The movements were constant and characteristic, and his speech was slow and hesitating. His mental condition was unaffected.

Bower's second case presented all of the features of hereditary chorea, but there was an absence of a history of the disease in any branch of his family. The patient was thirty-two years of age, and there were twitchings and irregular movements beginning at twenty-nine. He had no evidences of mental disturbance. The third was a woman, forty-one years of age, in whom choreic movements began at the age of thirty-six. Her father had the same affection, and what is very interesting, her baby of four months had almost constant choreic twitchings of both hands. In this patient there was slight beginning dementia. All of these patients were in the wards of the Philadelphia Hospital.

Korniloff, a Russian, has reported a case of hereditary[2] chorea. Hay, in a valuable article,[3] adds to the literature of the subject six cases which have come under his observation in the State Asylum for the Insane, Morris Plains, N. J., and gives notes of two more cases which were in

[1] Journal of Nervous and Mental Diseases, March, 1890, p. 131.
[2] Annual of the Universal Medical Sciences, 1890, vol. ii., c. 54.
[3] University Medical Magazine, June, 1890.

the same institution a few years previous, making eight additional cases in all. All of these patients were taken to the Asylum for mental diseases. It is singular that so few cases of this disease have been reported from other hospitals for the insane. In Diller's cases, referred to above, there were but five cases of the hereditary form of chorea among the thirty-nine adult choreas which were found among the inmates of twenty-nine asylums. Hay's cases are all well-marked examples of the disease under consideration, and all of them had a family history of chorea going back one or more generations. In one patient the choreic movements did not become constant until nearly her sixtieth year, although there had been several brief attacks previously. Another case became choreic at fifty-three years of age. The daughter of one of these cases became choreic before the age of twenty. In two of Hay's patients there was a history of acute rheumatism, and in two others, in which there was no record of rheumatism, there were cardiac murmurs.

Clarence King [1] makes a third communication on the subject of hereditary chorea, and gives the histories of two additional cases, a brother and a sister; both were insane, and were inmates of the Cattaraugus County Insane Asylum, N. Y. The maternal grandmother was the first among the ancestors in whom the disease could be traced. Mrs. S——, one of the cases, had the disease since she was twenty years of age, and was in the ward for imbeciles. Her brother, Henry C——, was sixty years of age, and had been choreic for fourteen years. Two of his children were said to have unmistakable signs of the disease, their ages being eighteen and sixteen years.

Dr. William Osler [2] reports the case of a man, aged fifty-nine years, who had had chorea for eight years. He had general irregular movements and his gait was peculiar.

[1] Philadelphia Medical News, July 15, 1890.

[2] Johns Hopkins Hospital Bulletin, December, 1890, vol. i., p. 110.

He walked with steps of irregular length, halting occasionally, and with stiffness of the legs. The reflexes were increased and there was slight ankle clonus. Four or five other members of the family were similarly affected. The expression was fatuous, and the mental condition was considered weak.

Arostegui[1] reports two cases. The details of these are not given in the abstract, which is the only report to which I have had access.

Biernacki[2] describes the case of a man, of forty-eight, who had been suffering from choreic movements for five years. His mother and maternal grandfather had had the same affection. His memory was considerably impaired.

Jolly[3] reports the following case: "Woman, thirty-five years old. Disease began at twenty-seven years. Patient gives as cause death of husband. First symptoms said to be twitching of face, but may have begun in leg. Finally the twitchings extended throughout the entire body. At the present time the rather anæmic woman presents the appearance of a rather severe case of ordinary chorea minor. The muscles of the face, trunk, and extremities are in constant movement. The brows are raised, mouth grins, eyes roll, and tongue protrudes in quick succession. The gait is rocking, the steps dancing and scraping, and sometimes, on account of sudden motion of the trunk or strong flying out of a foot, the gait is interrupted. The speech is often cut short by the smacking of the lips or sudden inspiration, as in many cases of acute chorea, while no true disturbance of articulation exists.

"The patient can, when comfortably quiet, write her name and short sentences. During sleep the motions cease. Mental disturbances of two kinds are recognizable. She herself complains of loss of memory, which can

[1] Crónica Médico-Quirúrgica de la Habana, from Annual of the Medical Sciences, 1891, vol. ii., c. 52.

[2] Berlin. klin. Wochen., 1890, xxvii., 485, and Annual of the Universal Medical Sciences, 1891, vol. xi., c. 51.

[3] Neurolog. Centralblatt, June 1, 1891.

also be objectively proved. The statements as to the history of her disease are unsafe and incomplete, and sometimes fabulous. She is very weak in reckoning, while formerly she was quick and intelligent. For the rest, she is by no means to be regarded as imbecile. The second mental disturbance concerns the emotional sphere. She is very irritable, easily led to tears, and often much depressed. At these times the choreic movements increase. Her previous temperament is said to have been quiet. The mother of the patient and two brothers of the mother suffered from the same disease. The mother became ill, in her thirty fifth year, in the same manner as the patient, and suffered till her death, aged forty-six years. In her, also, loss of memory was marked. In her later life she was feeble-minded, and at times maniacal. An elder brother of the mother became ill later than she, eight years before his death, from chronic chorea. He became imbecile. A younger brother of the mother will be exhibited by Dr. Remak. In this patient the chorea is less than in ——, and the mental affection is also less. Four brothers and sisters of the mother remain healthy. The patient has two sisters, aged thirty and twenty-five years, of whom the last suffers from migraine. In her generation she is the only one affected with chorea. Of the two living children of the patient, girls of ten and eleven years, the elder has suffered from chorea for two years, and is said to have had epileptic attacks for the same length of time."

I have had under my care, in the nervous wards of the Philadelphia Hospital, in the past two years, three patients with hereditary chorea, whose cases have not yet been reported. I am indebted to Dr. Alexander and Dr. John H. Rhein, internes of the Philadelphia Hospital, for the histories of these cases.

Case I.—William D——, aged sixty-three, pedler by occupation. Admitted to the nervous wards of the Philadelphia Hospital, July 16, 1889. The patient knows of no disease in the family except the one with which he

is affected. He states that his mother had the same affection. He thinks that in her case it came on late in life, and he knows that it continued until her death, which was at the age of about seventy. In his mother's case there was a general tremor, not so irregular, he says, or extreme, as in his own case. He knows of no other member of his family being affected with any nervous disorder. Patient has always had good health. He has not been intemperate or addicted to excesses of any kind. About fifteen years ago, that is, at the age of forty-eight, choreic movements began in his hands. There was at this time no cause to excite this condition. The movements gradually increased in extent, and by degrees involved the whole body, until the face and trunk became affected. About five years after the beginning of the trouble the irregular movements had extended to the whole body. He thinks that his disorder has been aggravated by treatment by arsenic.

Present Condition.—All of the muscles of the body, including the facial muscles, are involved in the choreic movements. There is continual twitching and jerking, and the movements are greatly exaggerated by any excitement. A strong effort of the will can control the movements for a short time, and during sleep they cease entirely. Speech is somewhat affected, his articulation being slow and halting. The knee-jerks are exaggerated, and all of the cutaneous reflexes are also increased. His mind is somewhat affected. He is not clear in giving an account of himself, but it is difficult to say whether he has any delusions. There is no pain, and no impairment of sensation. There are no bladder or rectal symptoms. His general health is good, and no disease of any of the abdominal or thoracic organs can be detected.

Case II.—Mary D——, aged forty-six, white; born in Ireland; occupation, housework. Admitted to the Philadelphia Hospital, July 20, 1890. Her grandparents died of old age. Mother died at the age of thirty, of con

sumption, according to the patient's statement. Five years ago her father had a stroke of paralysis. This was very slight, and kept him in bed only three weeks. He now walks well with the aid of a cane. His speech is slightly affected. She has two brothers, who are well and strong; one sister, who is dead, from causes unknown to patient. She says that her entire family is nervous, that is, all have slight tremor of the hands in holding the paper to read. She has had the diseases of childhood and small-pox; with these exceptions has always been healthy. She has never had rheumatism. Her menstrual flow was established at the age of fifteen; she has always been regular and there has been no dysmenorrhœa. She still menstruates regularly. She has always lived with her father, who has abused her constantly. She says, "he made a foot ball of her head." Two years ago he struck her an unusually hard blow on the side of her head. This hurt her very much at the time, but did not render her unconscious. A day or two after this she was asked by a neighbor why her head shook so much. She did not know that there were any movements. She soon found that her hands and the whole body were in constant motion. These movements do not tire her, and she is not conscious that they are made unless her attention is called to it, or she happens to look at herself in the looking-glass. She can read without difficulty. Her head is held a little on the right side, and has a constant back-and-forward or lateral movement. There are slight spasmodic inco-ordinate movements of both hands and feet, which can be arrested by attention or voluntary effort. The tremor is irregular and well-marked, without being coarse. Her gait is characteristic. She is able to walk, but after taking five or six steps in a deliberate fashion, there comes a long stride with one foot, then she brings up the other in a studied and measured way, much like a dancing step—then continues regular steps for five or six more, and again repeats the movement of bringing the second foot up, after a long stride. She cannot walk in

a straight line, going constantly off to one side or the other. All voluntary efforts are slow and labored. The tongue is protruded well, but is not held out for any time, it being returned suddenly, and apparently involuntarily. Choreic movements affect the muscles of the neck. The head is constantly being rotated to the left and upward, and there is apparently some contraction of the sterno-cleido-mastoid muscle which holds the head even when not moving in the above-described position. The intellect is impaired. The patient is voluble, and often repeats pointless remarks. She is easily excited, and one day, after having been alarmed at the prospect of an electrical examination, she was much excited, talked loudly, acted strangely, and was very restless for two hours. The pupils are small, equal, and responsive to light. There is slight lateral nystagmus. The reflexes are everywhere exaggerated; sensation is everywhere good, and there is no loss in motor power. Her lungs and heart are normal, and her general health is fairly good.

Examination, July 17, 1891.—Patient's general condition is about the same as in the last note. The choreic movements are more extensive. The eyelids cannot be kept quiet, and the orbicularis oris is in constant motion. The movements of the arms are peculiar. The left arm is flexed at an obtuse angle, with the hand extended to a right angle, with the arm and the fingers partially flexed. This position is constant, the movements exaggerating the position. The forearm is also rotated, and the whole arm moved upward and a little outward. The right arm is usually extended, and the movements on this side are more typically choreic, that is, the arm is not held stiff. The body rotates on the pelvis. The legs are also affected, the movements consisting of flexing of the feet and rotation of the legs, the thighs being occasionally flexed a little. The knee-jerk is now sluggish, elbow-jerks normal. The plantar reflex is very active. The gait is as before described, all the choreic movements are

exaggerated when she walks and cease during sleep. She complains of pains.

CASE III.—John W——, aged forty-eight, white; native of Germany; occupation, carpet-weaver. Admitted to the Philadelphia Hospital, July 2, 1890. His paternal grandfather is said to have had the same form of chorea as the patient, but no data can be obtained in regard to his case. The patient's father began to show irregular spasmodic movements of the head and extremities when he was forty years of age. He grew progressively worse and died at the age of fifty years. One sister began to show symptoms of the same disease at the age of fifteen. She is still living, but is in Germany, and the patient cannot tell what her present condition is as he has not seen her for many years. When twenty-nine years of age the patient first noticed a stiffness in his arms and hands, so marked that he could not work with his usual ease. Soon after this he found that his hands were beginning to move about in an irregular manner. He did not feel the movements himself, and would not have known that they were made unless he was looking at his hands. A year later the head became involved, and in a short time the movements extended to his legs. He has not been able to do work of any kind since thirty years of age.

Present Condition.—The patient is entirely unconscious of his movements. They do not cause him any pain, weariness, or inconvenience. The arms, head, and trunk are in continual movement, the movements being greatest if he is excited in any way; for instance, asking him questions will increase all of the movements to a marked degree. The movements are greater on the left side. He can feed himself, but with very great difficulty, and attempts at voluntary movements increase the chorea. His gait is peculiar and characteristic of the disease. It is very similar to the walk of Mary D——. In walking his whole attention seems absorbed by the effort. The step is measured and slow. The leg and thigh are first well flexed, then the weight of the body is slowly and

cautiously thrown on one foot, the same movements being repeated on the other side of the body. After six or eight steps have been taken in regular succession, one foot is suddenly thrown forward, making a long stride, then the other foot is brought up to a short distance from the rear of the first. The regular steps are then taken for the usual time, to be again followed by the long strides. These long strides usually alternate, and very seldom do two occur on the same side in succession. The whole walk looks studied and much like a dancing figure. The tongue cannot be kept protruded. There is no loss of power and there is no loss of sensation. Reflexes: The knee-jerk and all of the other reflexes are increased. Ankle clonus can be developed. The plantar, epigastric, and cremasteric reflexes are exaggerated, as are the arm tendon taps. The pupils are equal and respond to light. His memory is considerably impaired, he is reticent, and never converses or reads. His speech is jerky and unintelligible. He has exhibited no delusions but his mental calibre is evidently far below normal. His appetite is good. There is marked constipation, but his general health is excellent. Although uncommunicative and having but little intercourse with the other patients he was cheerful enough, and when asked how he was, always declared that he was getting better. In March, 1891, he fell while going down-stairs and fractured the neck of the right humerus; although carefully dressed the bone did not unite. His condition remained about the same until May 14, 1891. On this day while sitting in the ward he fell from the chair to the floor, being apparently thrown by one of his choreic movements. He was able to get up, and with some assistance was got into bed. The next morning he was in a semi-conscious condition. There was no paralysis, no change in the pupils, and by arousing him he apparently understood questions but did not reply to them. When told to do so he put out his tongue, and if the arm were raised it would be held up for some minutes. He took

nourishment when it was given to him. The stupor gradually became more profound and the patient died May 18th. From the time of the last fall until death the choreic movements ceased entirely. It was impossible to get the consent of the friends to make an autopsy.

These additional cases of hereditary chorea all bear a striking resemblance in their clinical features to those already published, and we are enabled by the increased material at our command to draw more definite conclusions than formerly.

1st. The fact of heredity is sustained. In one of Bower's cases, there was no history of the disease among the progenitors of the patient, but all of the symptoms were identical with those usually met with in hereditary chorea, and, as Dr. Mills justly remarked, "the disease must have a beginning," and this patient was probably the original case in his family. In one of my cases (Case II.) there is no clear history of heredity, but the symptoms are so typical that I think I am justified in classing it as a case of this disease.

2d. In most of the cases the disease began between the ages of thirty-five and fifty; but there are a number of exceptions to this rule. In one of Hay's cases the disease was not established until after sixty years of age, and in another, not until after the fiftieth year. In one of my last cases (Case III.), the irregular movements began at about twenty-nine years. Hoffman relates a case which began at ten years, and one who became affected while going to school. One of Lannois's cases developed the disease at twenty-one, and another, of Hay's, in youth. In King's last series of cases there are several in whom the affections began under twenty years of age. Mrs. S—— suffered from chorea for twenty years. Two of the children of Henry C——, the brother of Mrs. S——, were choreic at the age of sixteen and eighteen years. Suckling's patient had a daughter who was twelve years of age, and who showed, according to the author, unmistakable signs of the disease. Klippel's pa-

tient became choreic at fifteen years. A sister of one of my patients (Case III.) is said to have become choreic at fifteen years.

Mental disease occurs in most patients who are affected with hereditary chorea. In some the mental disturbance does not begin until the individual has been choreic for several years, but in others the insanity and chorea begin at the same time. Sometimes the want of mental equilibrium is noted first. The form of mental symptoms is generally of the same type. There is melancholia, with a tendency to suicide, irritability of temper, supposed delusions, and occasional outbreaks of violence are common. In the patients who have come under my own observation mental disturbance has been present, but has not been great. John W—— (Case III.) was melancholic, but had no delusions; and Mary D—— (Case II.) had mild dementia. In Huntington's patients, the two brothers, whose cases he related, there were delusions of grandeur; they thought they were captivating to the opposite sex, and made themselves conspicuous by their actions when in the street. This form of mental disorder is exceptional.

From a study of the cases now on record I have come to the conclusion that there are two forms of hereditary chorea, one in which the irregular muscular movements begin first, and, after a lapse of years, mental deterioration begins, and the other, in which the mental disease begins before, or simultaneously with, the chorea. The tendency to early or late development of insanity belongs to certain families. Rheumatism has occurred in a number of cases, notably in two of Hay's, and several patients have had valvular disease of the heart. The knee jerks are exaggerated in most of the cases in which the reflexes have been observed, and occasionally there has been recorded the presence of ankle clonus. King mentions a peculiarity in some of his patients which I have observed in two or three of mine. It is a tendency to stand in a certain position for long periods. For ex-

ample, Mrs. S——, one of King's patients, was accustomed to stand for many hours, especially when trying to masticate food, with her body thrown forward to a level of her hips, and her arms extended backward by the side of the body, the palms looking upward. One of my patients, David M——, would stand leaning against the side of a building for long intervals. The gait in this form of chorea deserves especial attention, for it is so unlike that of Sydenham's chorea. In several of the cases that I have seen the walk was much alike. The description of it in the case of John W—— (Case III.) is quite accurate. It is somewhat like a dancing step, measured and studied, but with a sudden and quick change in the time. There is apparently little change in the sexual function; several patients are mentioned as having borne children after becoming choreic. The pathology is still undetermined, but there is no doubt that the disease is one of developmental origin.

I have had the opportunity of making an autopsy in one case of hereditary chorea. The patient was David M——, whose case I reported in 1888.[1] He had suffered from the affection for ten years, and his death was due to exhaustion. He was worn out by the excessive movements, and during the last six months of his life gradually lost strength, and was finally confined to his bed, and died as a result of exhaustion due to the choreic movements, February 5, 1891. The skull-cap was extremely thick and the diploë was absent; the dura was adherent in places and was thick and tough; the meninges were congested and there was considerable œdema; the brain and cord were removed together. The membranes of the cord were congested, and to a small extent were adherent. There was no gross lesion observed, but the brain was not cut for the reason that it was preserved for later examination. The kidneys weighed about three and a half ounces each; the liver two pounds and five ounces. Though there was some slight sclerosis of the organs, there was no pro-

[1] Journal of Nervous and Mental Diseases, February, 1889.

nounced lesion anywhere found. The brain, unfortunately, was preserved in chloride of zinc solution, and therefore was useless for microscopic examination. The cord, however, was well preserved in Muller's fluid, and through the kindness of Dr. William M. Gray, of Washington, I am able to present these beautiful sections. The sections were examined under the microscope from the lumbar, dorsal, and cervical cord, stained with borax-carmine, and by Weigert's method. Macroscopic sections treated by the latter method show the following: All take the blue-black stain well in the posterior columns, while the remainder of the white matter stains poorly, especially in the dorsal and cervical regions. In the carmine sections the antero and lateral white matter stains too deeply, this again being most marked in the dorsal and cervical regions. The region of the central canal also stains a deep red. The cross sections are symmetrical in outline. Delineation of white and gray matter good. Microscopically the posterior columns are normal, except possibly a very slight increase of connective tissue in spots. In the remainder of the white matter there is considerable increase in the connective tissue. The walls of the blood-vessels are thickened, many axis-cylinders are missing, but never enough to completely cut off the tract. Motor-cells and nerve-roots normal. The region of the central canal is largely occupied by a mass of nuclear tissue similar to that found in normal cords, but the mass is far larger than usual, and stains much more deeply in the cervical region. The canal itself is quite large and irregular in outline. In the dorsal and lumbar regions it is small, and in one section shows as two canals.

Whether the condition described above be developmental, or whether it is due to a slow inflammation or to other causes, we are unable to say. The important change seems to us to consist rather in that which has taken place in the anterior and lateral white matter than that of the central canal.

There are many points of similarity between this affection and hereditary ataxia, which is now well established as a disease due to defective development of the nervous system. A striking feature in the histories of all of these cases is the marked neurotic taint through their families. Insanity, epilepsy, intemperance, and vice are prevalent in the family history. Hay calls attention to the frequent association of phthisis in the family. King refers to a circumstance to which I also alluded in my previous paper, namely, the probability that many, if not most, of the cases reported in this country are of common ancestry. East Hampton, L. I., according to King, was settled in 1649, and choreic families have lived there ever since. Several emigrations have taken place from there; one of them to Delaware County, N. Y., and it was from Franklin, Delaware County, that Dr. Waters's original observations were made. Charles B. Gorman, in his thesis, quoted by Dunglison, speaks of the affection existing in Luzerne County, Pa., which is less than seventy-five miles from Delaware County, N. Y. One of my cases was from Wyoming County, Pa., which adjoins Luzerne.

In closing, I would express the following conclusions: Hereditary chorea, while resembling in many respects Sydenham's chorea, differs in so many of its features that it is essentially a distinct and separate affection; that while, as a rule, there is remarkable uniformity in the symptoms presented, there may be variations; for example, in the occurrence of the disease at or before puberty. That it is not an invariable rule that if the disease fails to appear in one branch of the family the descendants of that branch have immunity. That the arrest of the movements by voluntary effort is not a distinguishing feature of hereditary chorea, as in some cases voluntary effort aggravates the movements, and there are many cases of Sydenham's chorea in which voluntary effort arrests the movements for the time. That chorea among the adult insane is a different affection from hereditary cho-

rea with insanity. That the evidence we have indicates that the pathology of the disease is a degeneration of imperfectly developed cells in the motor tract or in the cerebral cortex and in the spinal cord. The occurrence of the disease at an early age in children of some of the cases recorded is confirmatory of this view.

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